Physical Activity Level and Perception of Exercise in Cystic Fibrosis

David M Burnett, Ashley N Barry, and Joel D Mermis

BACKGROUND: More patients with cystic fibrosis (CF) are living longer, and lifestyle-related behavior is becoming increasingly important for improving morbidity and mortality. Declining levels of exercise leads to low cardiorespiratory fitness, which is a strong, independent predictor of mortality in patients with CF. As a result, exercise training has become a commonly accepted form of treatment for patients with CF. The purpose of this study was to determine physical activity levels and perception of exercise in adult patients with CF. METHODS: Adult patients from an in-patient CF unit were recruited to participate. A structured interview and self-report questionnaires were used to collect information on levels of physical activity and exercise perception including preferences, readiness, and barriers. RESULTS: Forty-six adult patients with CF consented to participate in the interview and completed self-report questionnaires. Subjects self-reported that the majority (84%) of their time was spent performing physical activity at a moderate level, with mean \pm SD of 11.8 \pm 12.2 h per week of moderate physical activity. Vigorous physical activity was described as hard and very hard physical activity, with a mean \pm SD of 1.8 \pm 4.6 h (13%) and 0.4 ± 1.6 h (3%), respectively. Most of the adult subjects with CF preferred walking, and 65% of them felt that exercise was very important. Lack of energy, lack of good health, lack of selfdiscipline, and lack of time were noted as the most frequent barriers to exercise. CONCLUSION: In this study, adult subjects with CF self-reported performing an adequate amount of moderate physical activity, although only a small proportion of time was spent at a vigorous level of physical activity. Clinicians providing rehabilitation have an opportunity to improve adherence to prescribed exercise by understanding the impact that physiological and psychological factors have on patients with CF. Further, motivating patients with CF to engage in more vigorous physical activity may provide a stimulus that improves clinical outcomes and potentially survival. Key words: cystic fibrosis; physical activity; exercise; aerobic fitness; exercise barriers; cystic fibrosis survival. [Respir Care 0;0(0):1-•. © 0 Daedalus Enterprises]

Introduction

Advances in treatment for cystic fibrosis (CF) are preserving lung function and increasing life expectancy. The proportion of adults in the overall CF population increased from 30% to 55% over the last 3 decades.¹ This trend is expected to continue, and the overall CF population is projected to increase by 50%, with a corresponding in-

The authors have disclosed no conflicts of interest.

crease of 75% in adult patients with CF.² As novel drug therapies continue to improve the life expectancy of patients with CF, there is an increasing emphasis on establishing strategies for improving symptomatic care and long-term health of adults with CF.

Although survival in CF is improving, new lifestyle challenges are emerging. It is unclear if patients with CF have adequate physical activity and exercise behavior pat-

The authors are affiliated with the University of Kansas Medical Center, Kansas City, Kansas.

This work was supported by the University of Kansas Medical Center's School of Health Professions.

Correspondence: David M Burnett PhD RRT AE-C, Respiratory Care and Diagnostic Science, Mailstop 1013, University of Kansas Medical Center, 3901 Rainbow Blvd, Kansas City, Kansas 66160. E-mail: dburnett@kumc.edu.

DOI: 10.4187/respcare.07193

terns to maintain a healthy cardiorespiratory fitness level. While patients with CF may perform a similar amount of physical activity as their non-CF peers, they perform significantly less vigorous physical activity (ie, exercise).³ Further, more time spent at a vigorous physical activity level is significantly related to increased cardiorespiratory fitness (r = 0.83).³ These results have clinical implications in that cardiorespiratory fitness is inversely associated with mortality^{4,5} and is one of the strongest predictors of survival in patients with CF.6 Accordingly, studies have reported that participation in exercise training programs and increasing physical activity levels have a positive impact on pulmonary function, cardiorespiratory fitness, fat-free mass, and quality of life for patients with CF.7-10 Based on the growing body of evidence related to exercise and CF, clinicians consider exercise as an accepted therapeutic strategy for improving the health of patients with CF.¹¹⁻¹³

To promote exercise as a component of the treatment plan for CF, a better understanding of how to improve adherence to exercise is needed to develop sustainable lifestyle change. Adherence to medical treatment has been a longstanding challenge in chronic illness, with reports of patient adherence ranging widely from 30-70%.¹⁴ Similar patterns of adherence to treatments are noted in patients with CF.¹⁵⁻¹⁹ Adherence varies within moderate to high ranges across treatment types in the population of adults with CF, including oral antibiotics (83%), pancreatic enzymes (85%), and bronchodilators (69%).¹⁸ In contrast, adherence to other CF treatments, including dietary changes and exercise, is poor (40–55%).^{15,18}

Although exercise has become an important component of the overall treatment plan for CF, integrating regular exercise into the daily routine of patients with CF can be difficult. Even though a large sample of adult subjects with CF (n = 545) indicated that exercise was an important part of their treatment plan, only 24% reported that they always adhere to exercise.²⁰ It seems reasonable to postulate that demanding and complex treatment routines present challenges to the adult patient with CF. Indeed, subjects with CF have reported that being too busy and tired are reasons for their lack of adherence.²¹ In contrast, strategies for improving adherence to exercise in patients with CF include individualized training, exercise variety, and homebased programs.²² These exercise program characteristics are consistent with a statement issued by the American Thoracic Society suggesting they should be used in a pulmonary rehabilitation program aimed at improving symptoms, exercise tolerance, and quality of life.²³ We plan to develop feasible and effective exercise program strategies for patients with CF; however, prior to developing an interventional design we must have a better understanding of the current physical activity levels and the perception of exercise in adults with CF. Therefore, the purpose of this study was to describe the physical activity levels as well as

QUICK LOOK

Current knowledge

The increasing life expectancy of patients with cystic fibrosis (CF) requires strategies for improving their longterm health. Exercise training has become an accepted therapeutic strategy for improving the health of patients with CF. Various levels of exercise intensities are now being used to elicit improvements in cardiorespiratory fitness which has been shown to be associated with survival in patients with CF.

What this paper contributes to our knowledge

Although adult subjects with CF performed an adequate volume of physical activity, a small proportion of time was spent engaged in vigorous physical activity. Subjects with CF felt that exercise is very important, but both physical and psychological factors are reported as barriers to exercise. Further, barriers to vigorous physical activity varied among the subjects, indicating that individualized rehabilitation strategies are needed.

perceptions related to exercise in adult patients with CF to better inform the development of the best strategy for effective exercise interventions. A secondary aim was to assess the relationship between physical activity levels and exercise perception to improve adherence for sustainable interventions.

Methods

This cross-sectional, descriptive study was conducted between June and December 2018 in an in-patient CF unit at a hospital on the University of Kansas Medical Center campus. This study was approved by the institutional review board. Patients with CF > 18 y old and able to communicate in English were included in the study. All subjects were recruited during their stay in the in-patient CF unit for treatment related to a pulmonary exacerbation. After consenting to participate in the study, they completed questionnaires and a face-to-face interview with a study team member.

Physical activity levels were measured with the 7-Day Physical Activity Recall questionnaire during a structured interview. The questionnaire and interview provided a costeffective instrument for measuring physical activity and can be used in a routine clinical setting. The questionnaire was developed as a general-purpose measure of physical activity²⁴ and has been widely used in epidemiologic, clinical, and behavior change studies. Specific to CF, the questionnaire has been validated for measuring physical activ-

Copyright (C) 2020 Daedalus Enterprises ePub ahead of print papers have been peer-reviewed, accepted for publication, copy edited and proofread. However, this version may differ from the final published version in the online and print editions of RESPIRATORY CARE

ity in patients with CF.25 The 7-Day Physical Activity Recall questionnaire is a semi-structured patient interview to estimate overall physical activity including light, moderate, hard, and very hard intensity levels. Light activities were explained as deskwork, standing, and light housework. Work-related, household, and recreational activity that makes one feel the same as walking at a normal pace were categorized as moderate physical activity. Very hard physical activity was described as similar to what it feels like when running at a continuous pace. The hard category fell between moderate and very hard. We calculated time spent in the moderate, hard, and very hard physical activity levels. In addition, physical activity time reported at a hard and very hard level was expressed as vigorous physical activity. As previously mentioned, it is suggested that vigorous physical activity is a sufficient stimulus for promoting cardiorespiratory fitness.3 Time was multiplied by the respective metabolic equivalent task (MET) and then summed to calculate the total MET minutes per week (METs/min/week), as explained in detail by Sallis et al.²⁴ The interviewer normally guided subjects through the previous 7 d prior to the interview. For subjects who experienced a recent exacerbation and subsequent hospitalization, they were asked to recall a normal week of physical activity and exercise when they felt well.

Perception of exercise was evaluated by assessing exercise preferences, readiness, and barriers. Exercise preferences and readiness were assessed with a study report form developed by the study team. Exercise preferences were obtained by asking an open-ended question; "What are, or what would be your exercise preferences if you engaged in regular exercise and/or sports activities?" Subjects were also asked 4 questions relating to their readiness for exercise. All answers related to readiness were recorded as a number from 0 to 10, with 0 being "not at all" and 10 being "very." The 4 questions about readiness for exercise as recommended by the medical team were scaled from 1 to 10 and included readiness to exercise, importance of exercise, self-motivation, and confidence to exercise. Barriers to exercise were measured with the Barriers to Regular Physical Activity domain from the Twin City Walking Survey.²⁶ This survey includes psychosocial variables associated with physical activity and consists of 15 questions. Each question was scored on a 5-point Likert scale to express how often (never, rarely, sometimes, often, very often) subjects felt that the variable noted prevents them from getting regular physical activity.

Descriptive statistics using mean \pm SD were used to explain physical activity levels and exercise perception. Common phrases were assessed for exercise preferences. Spearman correlation coefficients were used to analyze bivariate associations between physical activity levels and exercise perceptions. When distributions of variables were not normal, we used the median and interquartile range Table 1. Participant Characteristics

Participants, n	46
Male sex, %	52
Age, mean \pm SD (range)	31 ± 10.6 y (20–70)
Race/Ethnicity, n	
White	42
Black or African American	3
Hispanic or Latino	1

(IQR) to summarize the distributions. All analyses were performed with SPSS 25 (IBM, Armonk, New York).

Results

Subject characteristics are presented in Table 1. Fiftythree patients from the CF in-patient unit were asked to participate in the study, and 46 (87%) consented. The mean \pm SD age was 31 \pm 10.6 y.

Data collected from the 7-Day Physical Activity Recall questionnaire included physical activity levels categorized as moderate and vigorous (ie, hard and very hard) (Table 2). Total time of reported physical activity level at moderate and above varied considerably, ranging from 0 h to 39.5 h per week, with a mean \pm SD of 14.0 \pm 13.0 h per week. Subjects spent most of their physical activity time at a moderate level, as shown in Figure 1. Mean \pm SD time spent performing moderate physical activity was 11.8 ± 12.2 h per week. Mean \pm SD spent performing hard physical activity was 1.8 ± 4.6 h per week, and time spent performing very hard physical activity was 0.4 ± 1.6 h per week. Because the distributions for time at all levels of physical activity were not normal, we used the median and interquartile range (IQR) to summarize the distributions. The median (IQR) time spent in moderate physical activity was 8 (1.75–18.5) h; the median (IQR) time spent in hard physical activity was 0 (0-1.5) h), and in very hard physical activity was 0 (0-0) h. Overall, 44 and 23 subjects reported time spent in moderate and vigorous physical activity levels, respectively. The time spent in moderate and vigorous (ie, hard and very hard) physical activity was summed and used to calculate METs/min/week. The volume of moderate and vigorous physical activity varied widely, ranging from 0 to 12,780 METs/min/week, with a mean \pm SD of 3,709 \pm 3,488 METs/min/week.

Subjects' perceptions of exercise included self-report on their preferences, readiness, and barriers. The most common exercise preference was walking, as noted by 28 subjects (Fig. 2). Subjects also preferred the following activities: weights (n = 13), cycling (n = 12), jogging/running (n = 12), and dancing (n = 6). Other activities recognized by ≤ 3 subjects included basketball, golf, workout videos, calisthenics, Pilates, CrossFit, elliptical, and hiking. The

*	
Physical activity level, h	
Moderate physical activity	11.8 ± 12.2
Vigorous	
Hard physical activity	1.8 ± 4.6
Very hard physical activity	0.4 ± 1.6
Exercise preferences, number of times reported	
Walking	28
Weights	13
Cycling	12
Jogging/running	12
Dancing	6
Exercise readiness*	
Ready	7.4 ± 2.4
Important	9.2 ± 1.5
Motivated	6.0 ± 2.7
Confident	7.2 ± 2.7
Exercise barriers†	
Self-conscious about looks when exercising	1.8 ± 1.0
Lack of interest in exercise	2.8 ± 1.2
Lack of self-discipline	2.9 ± 1.2
Lack of time	2.9 ± 1.3
Lack of energy	3.3 ± 1.2
Lack of company	2.1 ± 1.2
Lack of enjoyment from exercise	2.6 ± 1.3
Discouragement	2.2 ± 1.2
Lack of equipment	2.2 ± 1.4
Lack of good weather	2.3 ± 1.2
Lack of skills	1.8 ± 0.9
Lack of facilities or space	2.0 ± 1.3
Lack of knowledge on how to exercise	1.8 ± 1.1
Lack of good health	3.2 ± 1.2
Fear of injury	1.5 ± 0.8

Table 2. Exercise Levels and Perception

Data are presented as mean \pm SD or *n*.

* Exercise readiness was reported on a scale of 0–10, 0 being "not at all" and 10 being "very." † Exercise barriers were reported on a 5-point Likert scale (1 = never, 2 = rarely,

3 = sometimes, 4 = often, and 5 = very often) and expressed how often participants felt the

variable noted in each question prevents them from getting regular physical activity.

mean \pm SD score for each exercise readiness component included importance (9.2 \pm 1.5), readiness (7.4 \pm 2.4), confidence (7.2 \pm 2.7), and motivation (6.0 \pm 2.7) (Table 2). In addition, 30 of 46 (65%) subjects selected the highest value (ie, 10, very important) when asked, "How important is it to exercise as prescribed by your medical team?" The mean \pm SD score for the 4 most frequent barriers included lack of energy (3.3 \pm 1.2), lack of good health (3.2 \pm 1.8), lack of self-discipline (2.9 \pm 1.2), and lack of time (2.9 \pm 1.3). In contrast, the mean \pm SD for the 4 least frequent barriers included fear of injury (1.5 \pm 0.8), lack of skills (1.8 \pm 0.9), self-conscious about looks when exercising (1.8 \pm 1.0), and lack of knowledge on how to exercise (1.8 \pm 1.1).

The relationships between physical activity levels (ie, moderate and vigorous) and exercise perception were as-

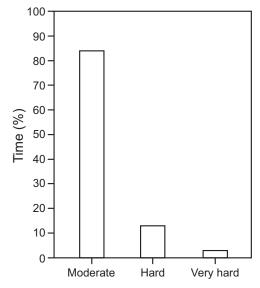


Fig. 1. Percent of time spent in moderate and vigorous (hard, very hard) physical activity levels.

sessed. There were significant correlations with moderate relationships between moderate physical activity with 3 of the 4 readiness for exercise components including readiness (r = 0.440, P < .002), motivation (r = 0.400, P < .002)P < .006), and confidence (r = 0.403, P < .006), but not importance. Also, moderate physical activity was significantly and negatively correlated with barriers to exercise. There were moderate, inverse relationships between moderate physical activity with lack of interest (r = -0.482, P = .001), lack self-discipline (r = -0.424, P < .003), and lack of good health (r = -0.591, P < .001). Further, there were weak, inverse relationships between moderate physical activity with lack of energy (r = -0.337, P = .02)and discouragement (r = -0.334, P = .02) There were no significant correlations between vigorous physical activity and exercise perception.

Discussion

Despite the benefits of physical activity, participation and adherence to routine vigorous physical activity are challenging. What level of physical activity do patients with CF engage in, and how do they perceive exercise? To address this question, we interviewed subjects with CF about their physical activity level. We also asked them to report on their perception of exercise based on their preferences, readiness, and barriers. We then assessed whether physical activity level was associated with exercise perception.

Thirty-seven (80%) of the subjects reported performing $\geq 150 \text{ min/week}$ of moderate and above physical activity. Moreover, 40 (87%) of the subjects reported engaging in $\geq 500 \text{ METs/min/week}$ of physical activity at a moderate

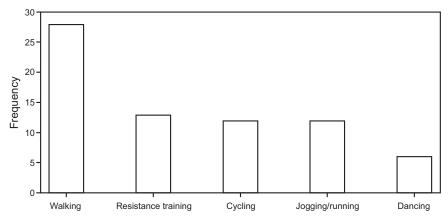


Fig. 2. Preferences for type of exercise are listed by their frequency as reported by all subjects.

level or above. Both of these results are in line with physical activity guidelines for healthy adults as suggested by the American College of Sports Medicine.²⁷ Although the source of physical activity volume is self-reported, it appears that a strong majority of our subjects are meeting physical activity guidelines for the general population.

With respect to the physical activity level, patients with CF may not feel confident in their ability to exercise at a level that makes them feel like they can't complete a full sentence while exercising (ie, at a vigorous level). Although we found that adults with CF engage in physical activity, a small proportion (16%) of the time was directed toward vigorous physical activity above a moderate level. A study by Currie et al²⁸ indicated that 38% of time spent during overall physical activity was associated with an intensity level above moderate. These results are consistent with another study reporting on vigorous physical activity in CF. Nixon et al³ indicated that subjects with CF perform less vigorous physical activity than their peers without CF. Further, increasing time spent performing vigorous physical activity has been reported to be linked to improved cardiorespiratory fitness,3 which in turn is directly associated with survival in CF.4-6 A possible explanation for the lack of participation in vigorous physical activity may be due to both physiological (ie, metabolic) and psychological reasons. Vigorous activity results in increased strain on the cardiopulmonary and musculoskeletal systems, and it is plausible that the increased metabolic demand and subsequent increased work of breathing is uncomfortable when increasing one's exercise intensity. Further, the perceptions of young adult subjects with CF regarding their inability to perform vigorous physical activity may be influenced from previous learned behavior derived from their parents' belief that they are unable to do such activities, given evidence indicating that healthy children's perceptions about vigorous physical activity was significantly related to their parents beliefs about them.²⁹

Based on previous evidence suggesting a link between vigorous physical activity, aerobic fitness, and survival, clinicians should consider strategies for incorporating "exercise" above walking at a normal pace into pulmonary rehabilitation programs for patients with CF. At the time of this study, a large, international, multicenter randomized controlled trial using vigorous physical activity in CF was in progress.³⁰ Support for the notion of a physical activity level above moderate was found in a meta-analysis reporting that the greatest reduction in relative risk for all-cause mortality was the result of increased time spent performing vigorous versus moderate physical activity.³¹ In addition, previous research supports the view that exercise at a higher intensity is more effective than moderate physical activity. Specifically, subjects with CF reported that high-intensity interval exercise was more time efficient and less strenuous compared to standard, moderate intensity exercise.32 Moreover, high-intensity interval training was able to elicit greater improvements in sustained submaximal exercise.32 Another study in subjects with severe COPD reported significantly greater adherence to interval training versus continuous training.33 Collectively, these studies support the theory that exercise and rehabilitation programs integrating vigorous physical activity can lead to improved adherence, ability to cope with demands of daily activities, cardiorespiratory fitness, and survival. However, safety of the patient should be the primary concern. Therefore, adequate training of rehabilitation specialists and proper screening of patients should be a priority. Future research should examine different levels of physical activity that elicit the greatest improvement in clinical outcomes to develop pulmonary rehabilitation guidelines for patients with CF.

Very few studies report on exercise perception (ie, preferences, readiness, barriers) in adult subjects with CF. Most subjects in this study (61%) preferred walking, along with weight training, cycling, and jogging. Another study by

White et al²¹ reported that 72% of adult subjects with CF indicated that walking was their most common type of exercise. Our subjects' perceptions of exercise suggest that they consider exercise to be very important. This finding may be due to the current consensus that exercise is associated with a healthy lifestyle. Although physical activity is considered to be very important by patients with CF, barriers remain that hinder participation in vigorous physical activity. Barriers to exercise noted in this study included the lack of energy, lack of good health, lack of self-discipline, and lack of time. This is consistent with evidence in the literature indicating that subjects with CF report being too tired and busy as barriers for adherence to exercise.²¹ Further, emotional and psychological barriers are reported to deter people with chronic illness from participating in exercise.³⁴ The combined effect of low energy, poorer health, lack of self-discipline, and insufficient time may take a psychological and emotional toll on patients with CF. These personal challenges may contribute to a lack of desire to participate in vigorous physical activity compared to their non-CF peers.35

Subjects in this study reported that moderate physical activity was moderately associated with perception about exercise (ie, readiness, motivation, confidence). Furthermore, subjects implied that moderate physical activity may be considered favorably by patients with CF based on its relationship with perceived level of interest, self-discipline, energy, discouragement, and good health. However, vigorous physical activity did not show a significant relationship with any exercise perception variable. We can only speculate that there is a significant degree of variation in the perception of exercise among those who engage in vigorous physical activity. For maximizing fitness benefits, exercise and rehabilitation interventions should consider individualized strategies for changing behavior, especially when incorporating an exercise stimulus above moderate intensity.

As clinicians, it's important to consider what the past evidence suggests about the most effective exercise stimulus for improving clinical outcomes in patients with CF. Furthermore, we must understand the exercise preferences, readiness, and barriers of patients with CF to create successful pulmonary rehabilitation strategies. Effective interventions featuring motivational coaching, an understanding of time constraints, exercise variety, and individualized training may help improve our patients' success in reaching their exercise and overall health goals.

Limitations

Limitations of this study include the cross-sectional, descriptive design. Therefore, our findings do not imply cause-and-effect relationships between the physical activity levels of patients with CF and their perceptions of exercise. We used self-reporting from a structured interview to assess physical activity levels. Although a structured interview was used to gather detailed information on physical activity, the number of total subjects was still small. However, using a cost-effective instrument enabled us to capture 25% of the CF population who routinely visit the CF Care Center while they were admitted to the inpatient CF unit. Further, we asked subjects to recall a typical week prior to hospitalization. As a result, there may be a level of recall bias when reporting physical activity. This study used a non-standardized questionnaire for measuring exercise preferences and readiness. However, the brief query from this questionnaire connected well with the 7-Day Physical Activity Recall and reduced burden on the subjects. Subjects from this study were selected from one site and cannot be generalized to the entire CF population.

Conclusion

Results from this study suggest that subjects with CF perform an adequate volume of physical activity, although only a small proportion of time is devoted to vigorous physical activity. Subjects prefer walking as a form of physical activity and believe it is very important to perform exercise as prescribed by their medical team. Further, subjects felt ready, motivated, and confident to perform moderate physical activity described as walking at a normal pace. Most common barriers to exercise were lack of energy, good health, self-discipline, and time. Clinicians involved in pulmonary rehabilitation programs should apply both physiological principles and psychological strategies for coaching and encouraging vigorous physical activity as a stimulus for improving cardiorespiratory fitness in patients with CF. In turn, patients with CF may achieve an active lifestyle that promotes improved survival.

ACKNOWLEDGMENT

We thank Jianghua He PhD for her help with the statistical methods and analyses.

REFERENCES

- Cystic Fibrosis Foundation Patient Registry. Available at: https:// www.cff.org/research/researcher-resources/patient-registry. Accessed September 19, 2019.
- Burgel PR, Bellis G, Olesen HV, Viviani L, Zolin A, Blasi F, et al. Future trends in cystic fibrosis demography in 34 European countries. Eur Respir J 2015;46(1):133-141.
- Nixon PA, Orenstein DM, Kelsey SF. Habitual physical activity in children and adolescents with cystic fibrosis. Med Sci Sports Exerc 2001;33(1):30-35.
- Hebestreit H, Hulzebos EH, Schneiderman JE, Karila C, Boas SR, Kriemler S, et al. Cardiopulmonary exercise testing provides additional prognostic information in cystic fibrosis. Am J Respir Crit Care Med 2019;199(8):987-995.

Copyright (C) 2020 Daedalus Enterprises ePub ahead of print papers have been peer-reviewed, accepted for publication, copy edited and proofread. However, this version may differ from the final published version in the online and print editions of RESPIRATORY CARE

- Vendrusculo FM, Heinzmann-Filho JP, da Silva JS, Perez Ruiz M, Donadio MVF. Peak oxygen uptake and mortality in cystic fibrosis: systematic review and meta-analysis. Respir Care 2019;64(1):91-98.
- Nixon PA, Orenstein DM, Kelsey SF, Doershuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. New Engl J Med 1992;327(25):1785-1788.
- Moorcroft AJ, Dodd ME, Morris J, Webb AK. Individualised unsupervised exercise training in adults with cystic fibrosis: a 1 year randomised controlled trial. Thorax 2004;59(12):1074-1080.
- Orenstein DM, Franklin BA, Doershuk CF, Hellerstein HK, Germann KJ, Horowitz JG, et al. Exercise conditioning and cardiopulmonary fitness in cystic fibrosis: the effects of a three-month supervised running program. Chest 1981;80(4):392-398.
- Schneiderman JE, Wilkes DL, Atenafu EG, Nguyen T, Wells GD, Alarie N, et al. Longitudinal relationship between physical activity and lung health in patients with cystic fibrosis. Eur Respir J 2014; 43(3):817-823.
- Selvadurai HC, Blimkie CJ, Meyers N, Mellis CM, Cooper PJ, Van Asperen PP. Randomized controlled study of in-hospital exercise training programs in children with cystic fibrosis. Pediatr Pulmonol 2002;33(3):194-200.
- Dwyer TJ, Elkins MR, Bye PT. The role of exercise in maintaining health in cystic fibrosis. Curr Opin Pulm Med 2011;17(6):455-460.
- Radtke T, Nevitt SJ, Hebestreit H, Kriemler S. Physical exercise training for cystic fibrosis. Cochrane Database Syst Rev 2017;11: CD002768.
- Rand S, Prasad SA. Exercise as part of a cystic fibrosis therapeutic routine. Expert Rev Respir Med 2012;6(3):341-351.
- Meichenbaum D TD. Treatment adherence: terminology, incidence and conceptualization. New York: Plenum Press;1987:19-39.
- Abbott J, Dodd M, Bilton D, Webb AK. Treatment compliance in adults with cystic fibrosis. Thorax 1994;49(2):115-120.
- Abbott J, Dodd M, Webb AK. Health perceptions and treatment adherence in adults with cystic fibrosis. Thorax 1996;51(12):1233-1238.
- Arias Llorente RP, Bousono Garcia C, Diaz Martin JJ. Treatment compliance in children and adults with cystic fibrosis. J Cyst Fibros 2008;7(5):359-367.
- Conway SP, Pond MN, Hamnett T, Watson A. Compliance with treatment in adult patients with cystic fibrosis. Thorax 1996;51(1): 29-33.
- Kettler LJ, Sawyer SM, Winefield HR, Greville HW. Determinants of adherence in adults with cystic fibrosis. Thorax 2002;57(5):459-464.
- Myers LB. An exploratory study investigating factors associated with adherence to chest physiotherapy and exercise in adults with cystic fibrosis. J Cyst Fibros 2009;8(6):425-427.

- White D, Stiller K, Haensel N. Adherence of adult cystic fibrosis patients with airway clearance and exercise regimens. J Cyst Fibros 2007;6(3):163-170.
- Prasad SA, Cerny FJ. Factors that influence adherence to exercise and their effectiveness: application to cystic fibrosis. Pediatr Pulmonol 2002;34(1):66-72.
- 23. Spruit MA, Singh SJ, Garvey C, ZuWallack R, Nici L, Rochester C, et al. An official American Thoracic Society/European Respiratory Society statement: key concepts and advances in pulmonary rehabilitation. Am J Respir Crit Care Med 2013;188(8):e13-e64.
- Sallis JF, Haskell WL, Wood PD, Fortmann SP, Rogers T, Blair SN, et al. Physical activity assessment methodology in the Five-City Project. Am J Epidemiol 1985;121(1):91-106.
- Ruf KC, Fehn S, Bachmann M, Moeller A, Roth K, Kriemler S, et al. Validation of activity questionnaires in patients with cystic fibrosis by accelerometry and cycle ergometry. BMC Med Res Methodol 2012;12:43.
- Forsyth A, Oakes JM, Schmitz KH. Test-retest reliability of the Twin Cities Walking Survey. J Phys Act Health 2009;6(1):119-131.
- American College of Sports Medicine. Philadelphia: Lippincott Williams & Wilkins;2009.
- Currie S, Greenwood K, Weber L, Khakee H, Legasto M, Tullis E, et al. Physical activity levels in individuals with cystic fibrosisrelated diabetes. Physiother Can 2017;69(2):171-177.
- Kimiecik JC, Horn TS, Shurin CS. Relationships among children's beliefs, perceptions of their parents' beliefs, and their moderate-tovigorous physical activity. Res Q Exerc Sport 1996;67(3):324-336.
- Hebestreit H, Lands LC, Alarie N, Schaeff J, Karila C, Orenstein DM, et al. Effects of a partially supervised conditioning programme in cystic fibrosis: an international multi-centre randomised controlled trial (ACTIVATE-CF): study protocol. BMC Pulm Med 2018;18(1): 31.
- Samitz G, Egger M, Zwahlen M. Domains of physical activity and all-cause mortality: systematic review and dose-response meta-analysis of cohort studies. Int J Epidemiol 2011;40(5):1382-1400.
- Gruber W, Orenstein DM, Braumann KM, Beneke R. Interval exercise training in cystic fibrosis effects on exercise capacity in severely affected adults. J Cyst Fibros 2014;13(1):86-91.
- Puhan MA, Busching G, Schunemann HJ, VanOort E, Zaugg C, Frey M. Interval versus continuous high-intensity exercise in chronic obstructive pulmonary disease: a randomized trial. Ann Intern Med 2006;145(11):816-825.
- Rimmer JH, Riley B, Wang E, Rauworth A, Jurkowski J. Physical activity participation among persons with disabilities: barriers and facilitators. Am J Prev Med 2004;26(5):419-425.
- Marcus BH, Dubbert PM, Forsyth LH, McKenzie TL, Stone EJ, Dunn AL, et al. Physical activity behavior change: issues in adoption and maintenance. Health Psychol 2000;19(1s):32-41.

Respiratory Care $\bullet \bullet \bullet$ Vol \bullet No \bullet